

Primary myelofibrosis

- Myeloproliferative neoplasm in which there is the replacement of bone marrow with collagenous connective tissue and progressive fibrosis.
- Characterized by
 - Extramedullary hematopoiesis
 - Progressive splenomegaly
 - Anemia
 - Variable change in the number of granulocytes and platelets including thrombocytopenia

Myelofibrosis

- Chronic myeloproliferative disorder resulting in marrow fibrosis
- Most cases are **secondary** to other processes
 - Malignant: leukemia or lymphoma
 - Essential thrombocytopenia
 - Multiple nonmalignant etiologies
- **Primary myelofibrosis**: 2 classes
 - Agnogenic myeloid metaplasia with myelofibrosis: indolent myeloproliferative syndrome (usually displays splenomegaly)
 - •Acute myelofibrosis (usually no splenomegaly)

Imaging

■ Location

- Axial skeleton, including pelvis and shoulder girdles
- Long tubular bones, proximal > distal

■ Radiograph/CT

- Marrow osteosclerosis (or normal)
- Hepatosplenomegaly
- May have extramedullary hemopoiesis

■ MR

- T1: **very low signal (lower than disc or muscle)**
- Signal intensity (SI) remains low on T2 or STIR
- No enhancement of marrow
- Fat in marrow is replaced; opposed-phase imaging does not show ↓ in SI

Case 28
53 y/o male

Myelofibrosis

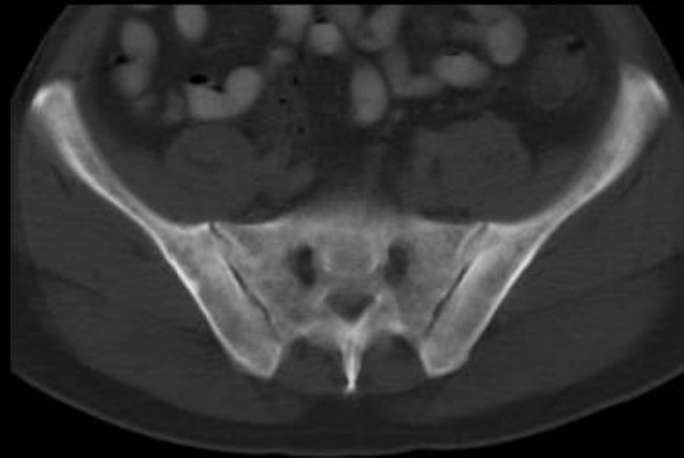
CT –Abdomen

Diffuse osteosclerosis of
sacrum and iliac bones without
architectural distortion

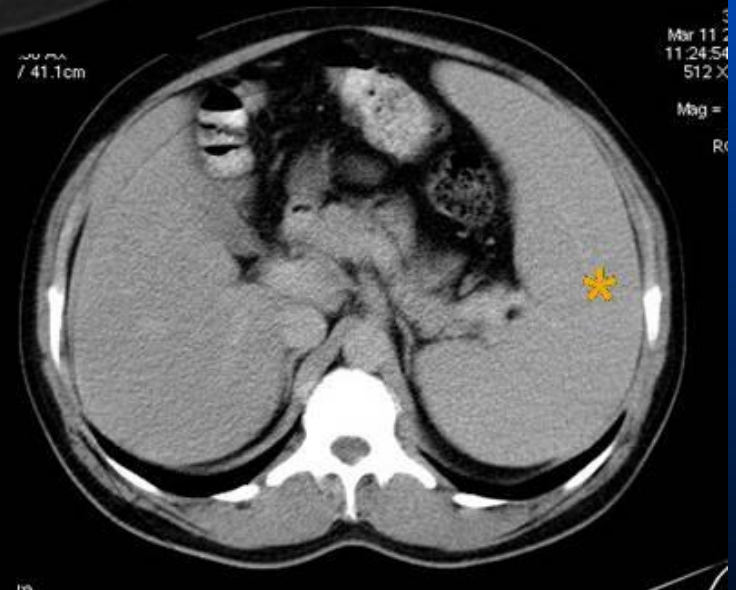
Splenomegaly (*)

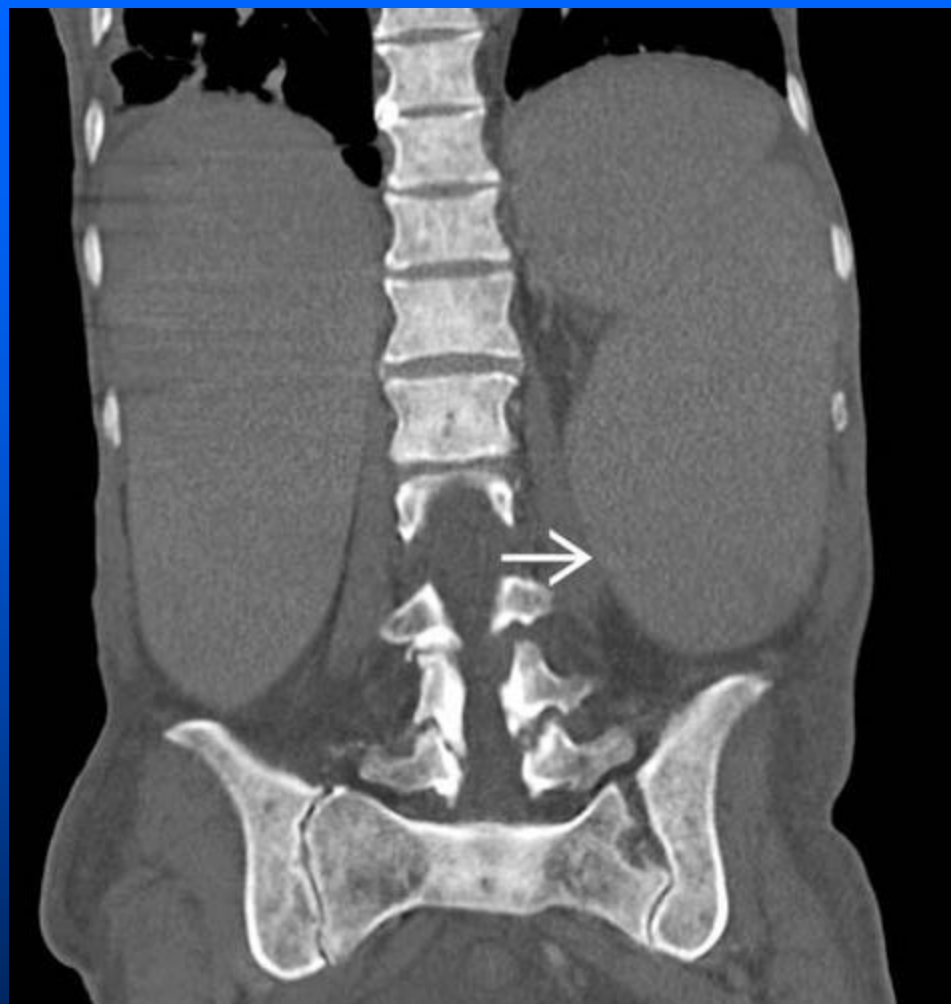
Myelofibrosis is a
haematological disorder where
there is replacement of bone
marrow with collagenous
connective tissue and
progressive fibrosis.

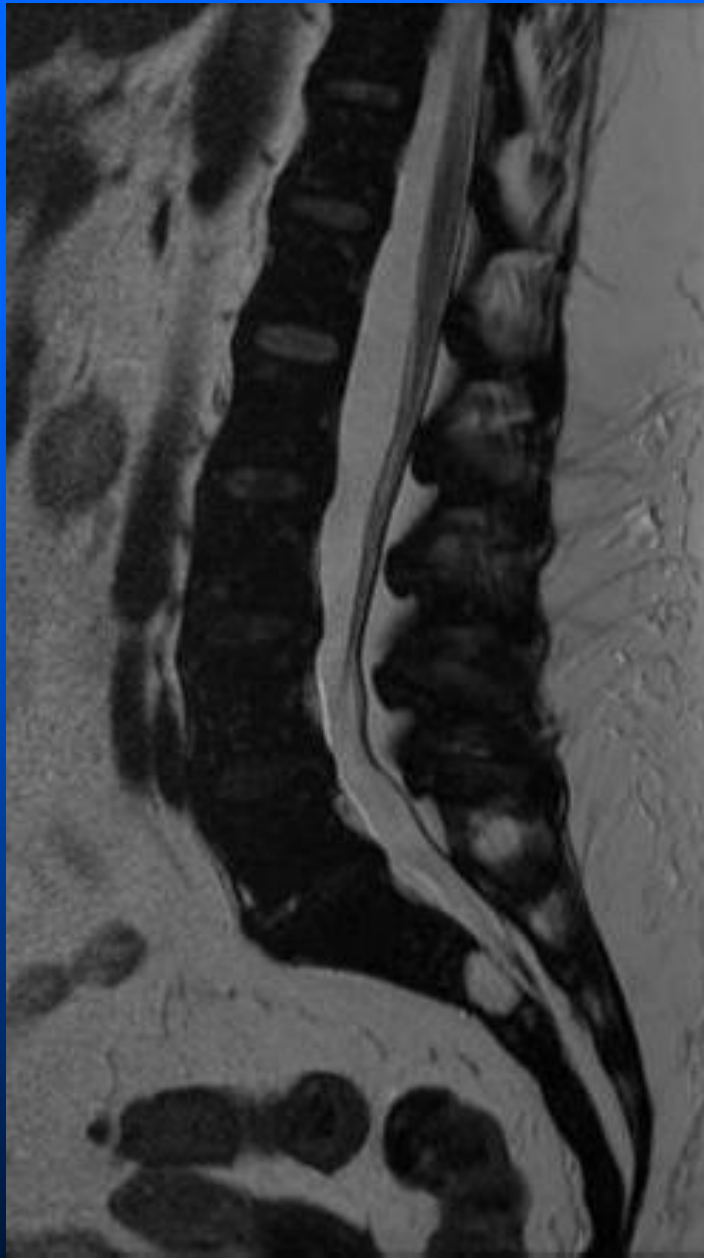
- Characteristic features:
Extramedullary hematopoiesis
- Progressive splenomegaly
- anemia
- variable change in number of
granulocytes and platelets

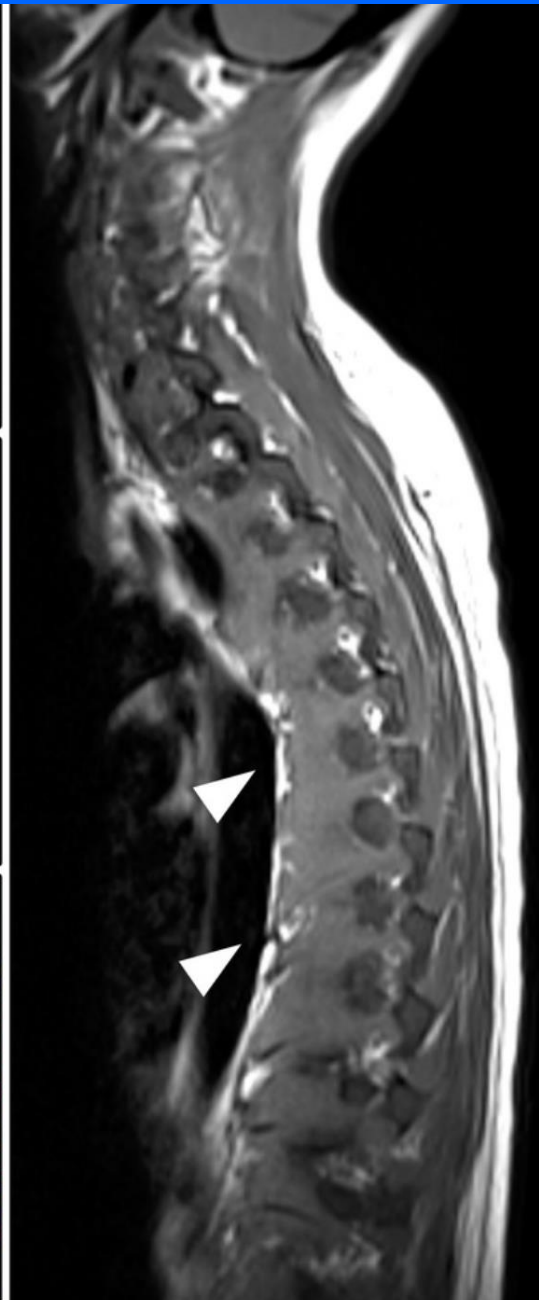
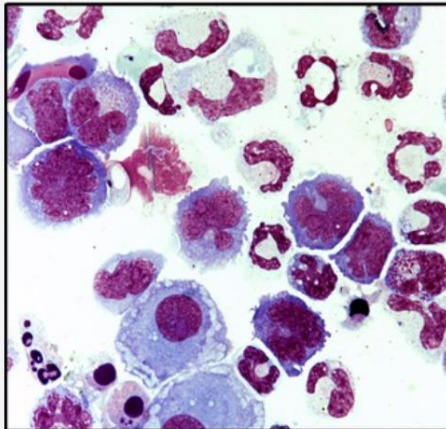
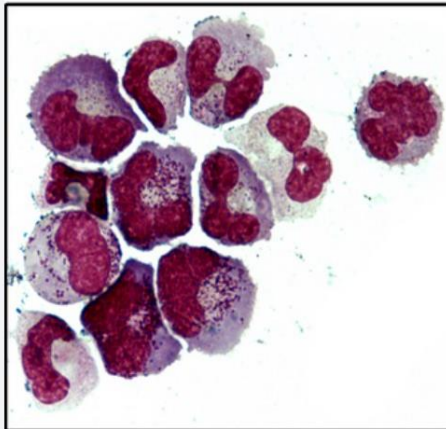
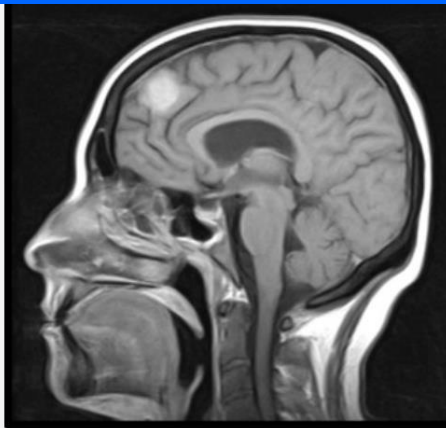


41.1cm









Non-hepatosplenic extramedullary haematopoiesis in primary myelofibrosis -